Journal of Neurosciences in Rural Practice
Home
Current issue
Instructions
Submit article

J Neurosci Rural Pract. 2015 Jul-Sep; 6(3): 405–406. PMCID: PMC4481799

Commentary

Valerie Purvin

Department of Ophthalmology and Neurology, Indiana University Medical Center, Indianapolis, Indiana

Address for correspondence: Dr. Valerie Purvin, Department of Ophthalmology and Neurology, Indiana University Medical Center, Indianapolis, Indiana. E-mail: vpurvin@iupui.edu

Mucormycosis is an opportunistic fungal infection that predominately affects immunocompromised patients, particularly those with diabetic ketoacidosis. The infection is typically rapidly progressive and potentially lethal due its predilection for spread along vessels with consequent thrombosis. This secondary infarction can affect the brain, causing stroke, and the orbit, producing visual loss, ophthalmoplegia and proptosis.

Infarction of the retrobulbar portion of the optic nerve is termed posterior ischemic optic neuropathy and is an uncommon event, particularly in contrast to ischemia of the optic disc.[1] The posterior portion of the optic nerve has a rich vascular supply from branches of the ophthalmic and retinal arteries with anastamoses between central and pial (peripheral) arteries, making it relatively immune to damage from vascular occlusion.

The three main settings in which we see posterior ischemic optic neuropathy (PION) are: In the perioperative period following prolonged surgery (usually spine or heart), secondary to giant cell arteritis, and with atherosclerotic risk factors.[2] Historically, the diagnosis of PION has been a clinical one, manifest as acute visual loss not explained by ocular pathology and with a normal appearing optic disc. With advances in neuroimaging we can now confirm and further expand on this diagnosis with MRI, particularly diffusion weighted imaging (DWI). Restricted diffusion on DWI, when combined with ADC mapping, is highly sensitive and specific for infarction and shows signal change within hours of onset of the ischemic event.

Restricted diffusion within the optic nerve was initially demonstrated in peri-operative PION[3] and subsequently in mucormycosis,[4] cavernous sinus thrombophlebitis,[5] thrombocytosis[6] and optic peri-neuritis.[7] Similar imaging findings in a case of lymphoma were attributed to hypercellularity due to tumor rather than infarction.[8] Infarction of the retina as well as the optic nerve has also been demonstrated in mucormycosis.[9]

DWI changes in the optic nerve in mucormycosis have been described within just a few hours of visual loss, at a time when no orbital signs were present, indicating the sensitivity of this modality.[10] Because the outcome in mucormycosis is highly dependent on prompt and aggressive treatment, any technique that allows for earlier diagnosis is valuable.

The diagnosis of rhinocerebral mucormycosis should be suspected in any immunosuppressed, debilitated or diabetic patient who develops facial or orbital pain, diplopia, orbital signs (proptosis, chemosis, lid

swelling) or acute optic neuropathy. The onset of acute unilateral or bilateral visual loss with normal optic discs should prompt neuroimaging, even without these specific risk factors. CT scanning of the orbit in mucormycosis with visual loss may demonstrate enlargement and enhancement of the optic nerve, which are non-specific. Routine MRI sequences will show similar non-specific changes. The presence of restricted diffusion on DWI sequences, however, is considered specific for infarction. In the absence of a history of hypotension with blood loss or signs and symptoms of giant cell arteritis, and in the presence of the above risk factors, a diagnosis of mucor is most likely. Definitive diagnosis usually rests on pathologic examination of affected tissue but treatment can be initiated while biopsy is pending. Correction of underlying metabolic abnormalities, when possible, also affects the prognosis for recovery.

References

1. Ghuman MS, Kaur S, Bhandal SK, Ahluwalia A, Saggar K. Bilateral Optic Nerve Infarction In Rhino-cerebral Mucormycosis. J Neurosci Rural Pract. 2015;6:403–4. [PMCID: PMC4481798]

2. Sadda SR, Nee M, Miller NR, Biousse V, Newman NJ, Kouzis A. Clinical spectrum of posterior ischemic optic neuropathy. Am J Ophthalmol. 2001;132:743–50. [PubMed: 11704036]

3. Purvin V, Kuzma B. Intraorbital optic nerve signal hyperintensity on magnetic resonance imaging sequences in perioperative hypotensive ischemic optic neuropathy. J Neuroophthalmol. 2005;25:202–4. [PubMed: 16148628]

4. Mathur S, Karimi A, Mafee MF. Acute optic nerve infarction demonstrated by diffusion-weighted imaging in a case of rhinocerebral mucormycosis. AJNR Am J Neuroradiol. 2007;28:489–90. [PubMed: 17353318]

5. Chen JS, Mukherjee P, Dillon WP, Wintermark M. Restricted diffusion in bilateral optic nerves and retinas as an indicator of venous ischemia caused by cavernous sinus thrombophlebitis. AJNR Am J Neuroradiol. 2006;27:1815–26. [PubMed: 17032847]

6. Klein JP, Cohen AB, Kimberly WT, Shah AS, Leiderman YI, Cestari DM, et al. Diffusion-weighted magnetic resonance imaging of bilateral simultaneous optic nerve infarctions. Arch Neurol. 2009;66:132–3. [PubMed: 19139313]

7. Spierer O, Ben Sira L, Leibovitch I, Kesler A. MRI demonstrates restricted diffusion in distal optic nerve in atypical optic neuritis. J Neuroophthalmol. 2010;20:31–3. [PubMed: 20182203]

8. Sudhakar P, Rodriguez FR, Trobe JD. MRI restricted diffusion in lymphomatous optic neuropathy. J Neuroophthalmol. 2011;31:306–9. [PubMed: 21623229]

9. Hatipoglu HG, Gurbuz MO, Yuksel E. Restricted diffusion in the optic nerve and retina demonstrated by MRI in rhino-orbital mucormycosis. J Neuroophthalmol. 2009;29:13–5. [PubMed: 19458569]

10. Alsuhaibani AH, Al-Thubaiti G, Al Badr FB. Optic nerve thickening and infarction as the first evidence of orbital involvement with mucormycosis. Middle East Afr J Ophthalmol. 2012;19:340–2. [PMCID: PMC3401808] [PubMed: 22837632]